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Case Report

The role in the decision-making of the surgeon in choledochal cysts during pregnancy

Carlos F. Nicolás-Cruz[1]*, Paola I. Pech-Cervantes[1], Juan A. Loyo-Cosme[1], Omar M. Herrera-Ortiz de Ora[1], Jorge A. Ordoñez-García[2], Fabiola Valdez-Maldonado[3], Jose Manuel Degante-Aguilar[4]

[1]Department of General Surgery, Zone General Hospital "Nueva Frontera", Mexican Social Security Institute. Chiapas, México
[2]Department of Obstetrics and Gynecology, [3]Department of Anesthesiology, [4]Hepato-Pancreato-Biliary Clinic. Regional High Specialty Hospital "Ciudad Salud", National System of High Specialty Hospitals. Chiapas, México

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***Correspondence:**
Dr. Carlos F Nicolas-Cruz,
E-mail: nicolascarlosf12@gmail.com

ABSTRACT

The diagnosis and management of bile duct diseases during pregnancy are highly controversial because preserving both the fetus and pregnancy often required surgical intervention. In this manuscript we reported three cases of pregnant patients referred to our hospital with choledochal cyst. Medical management was implemented until fetal well-being was compromise with moderate cholangitis and oligohydramnios. A C-section prior induction with lung ripeness was performed, four weeks later cyst resection and biliodigestive derivation was performed prior preoperative parenteral total nutrition was implemented. All patients and products survived without complications.

Keywords: Choledochal cyst, Pregnancy, Hepp couinaud biliary-digestive derivation

INTRODUCTION

Choledochal cysts are dilatations of different portions of the biliary tract. They account for approximately 1% of all benign biliary diseases and occur most frequently in the Asian population with 1:100,000 to 1:150, 000. Biliary cysts are more common in females with a female: male ratio of 3–4:1.1.[1]

Since 1977, five types of choledochal cyst have been differentiated according to Todani's Classification: type I is subdivided into cystic choledochal in a strict sense (Ia); segmental dilatation (Ib) and diffuse or cylindrical dilatation (Ic); type II supraduodenal diverticulum; type III choledochocele; type IV extrahepatic and intrahepatic spindle cysts (IVa) or extrahepatic multiple (IVb); and type V multiple intrahepatic saccular dilatations or Caroli's disease.[2] This entity represents an important surgical challenge with the most frequent occurrence in childhood, however, 20% is recognized in adults.[3] The classical clinical presentation is with right upper pain, palpable abdominal mass, and jaundice; still all clinical signs are observed only in 20%-30% of all patients, being the most common symptoms for adult patients: pain, fever and nausea.[4,5] Although clinical signs are not considered definitive during pregnancy; upper abdominal pain is the most frequent symptom in previously reported cases during pregnancy.[6]

The management of bile duct diseases during pregnancy is highly controversial, even more if is required surgical intervention to avoid complications associated with pancreaticobiliary reflux, preserving the fetal and pregnant physical wellbeing and safety.[7,8] In this article we presented the first case series of choledochal cysts described with a surgical perspective in Latin American population, highlighting the surgical, operational, and medical decision challenge it represents. All patients

were informed about the risks and possible complications of the surgical procedure. The patients in this article consent the procedure, pictures and participation in this series.

CASE REPORT

Case 1

A 20-year-old female with 3 months of moderate to intense pain in epigastric and right upper quadrant, treated empirically with oral nonsteroidal anti-inflammatory drugs (NSAIDs), with subsequent appearance of intermittent jaundice. An abdominal ultrasound was performed reporting a dilatation of the biliary duct with no clear delimitation of the hepatic an choledochal duct, then a cholangio-MRI was performed to evaluate the biliary duct, diagnosing choledochal cyst Todani type 1A (Figure 1) also is demonstrated incidentally a unique intrauterine live product of 31 weeks of gestation with a weight ap-proximate of 1721 grs, Bilirubin and liver function enzymes then were normal, however two days later there was a significant increase in liver and pancreatic function tests. A mild cholangitis was diagnosed, with intermittent course during the following four weeks. Inducers of pulmonary maturation were administered, and a C-section was performed at 35 weeks gestation, without complications. The Apgar score was 8/9. Four weeks later with a preoperative preparation including total parenteral nutrition for 5 days a cyst resection and biliodigestive derivation type Hepp Couinaud was done. it consists in an hepaticojejunal anastomosis end to side Roux-en- Y; finding a 15×10 cm choledochal cyst and gallbladder with normal characteristics. (Figure 2-5) No complications were presented during the intervention. Enteral nutrition started on the fifth day and the patient was sent home at the seventh day.

Table 1: Clinical features, choledochal cyst characteristics, pregnancy, and newborn outcomes.

Patient no. case and age	Clinical presentation	Type of choledochal cyst	Weeks of gestation at the diagnosis	Emergency surgical procedure	Reason of emergency surgical procedure	Weeks of pregnancy at the resolution and via	Weight and Newborn Apgar	Complication After definitive surgical treatment
Case 1 20-year-old	Intense pain in epigastric and right upper quadrant	Todani 1A	31 weeks	Not required	Mild cholangitis	35 weeks C- Section	2150 gr; 8/9	None
Case 2 16-year-old	Mild and diffuse abdominal pain	Todani 1B	21 weeks	Yes. laparotomy with drainage of biliary cyst and T-catheter placement	Moderate cholangitis	27 weeks C- Section	1730 gr 8/9	Premature and low birth weight
Case 3 19-year-old	Abdominal pain and jaundice,	Todani 1B	30 weeks	Yes C- Section	Cholangitis and oligohydramnios	35 weeks C- Section	2030 gr; 8/8	Newborn respiratory distress

Case 2

A 16-year-old pregnant female with 21 weeks of gestation, presents with abdominal pain, an abdominal ultrasound was performed reporting a choledochal cyst, then a Cholangio MRI was performed, reporting a Todani type 1B cyst, conservative management was indicated with broad spectrum antimicrobials; however, fever, jaundice and abdominal pain persist, diagnosing moderate cholangitis. An abdominal laparotomy with drainage of biliary cyst and T-catheter placement was performed at the 23 weeks of gestation. The patient was referred to the liver, biliary and pancreas clinic of this Hospital at 27 weeks of gestation, then was planned to achieve the pregnancy at term, however, intrauterine growth restriction was demonstrated, so in consensus with obstetrics and gynecology department was decided to perform a C-section to obtain the single live intrauterine product of 27 weeks without complications with 8/9 Apgar score. Four weeks later, complete resection of biliary cyst and biliodigestive diversion type Hepp Couinaud with hepatic jejunum and Roux-en- Y anastomosis was performed. Same preoperative and postoperative preparation scheme was implemented without complications.

Case 3

A 19-year-old pregnant female with 30 weeks of gestation, presents with abdominal pain and jaundice, she was admitted in a rural hospital where abdominal and obstetric ultrasound was performed reporting a biliary cyst.

A Cholangio MRI was performed diagnosing a choledochal cyst Todani type 1B, Conservative management was decided until reaching the highest fetal viability, however, during the 35th week of gestation, the patient presented recurrent cholangitis, in addition to oligohydramnios; a C-section was performed obtaining the product alive with Apgar score 8/8 and newborn respiratory distress syndrome requiring supplemental oxygen with cephalic helmet for 24 hours until spontaneous recovery. Then a complete resection of the biliary cyst and biliodigestive diversion type Hepp Coinaud with Roux-en-Y hepaticojejunal anastomosis was performed four weeks later.

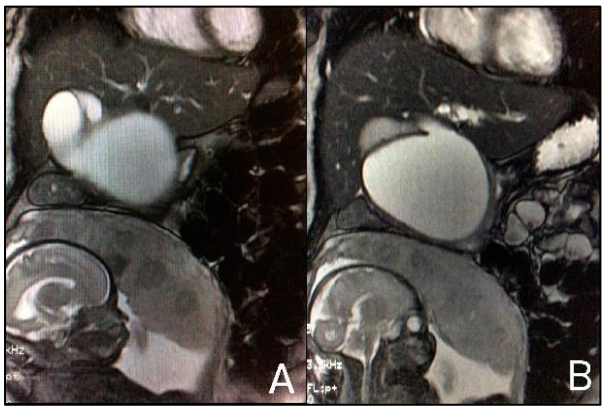

Figure 1: A) and B) Coronal view of magnetic resonance imaging showing the choledochal cyst and gravid uterus.

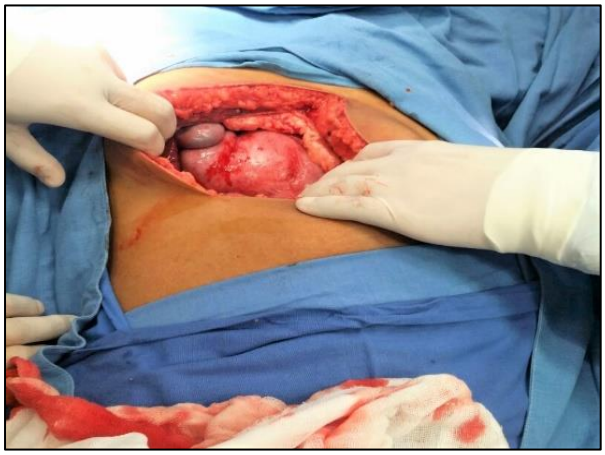

Figure 2: Surgical exposure of choledochal cyst and gallbladder.

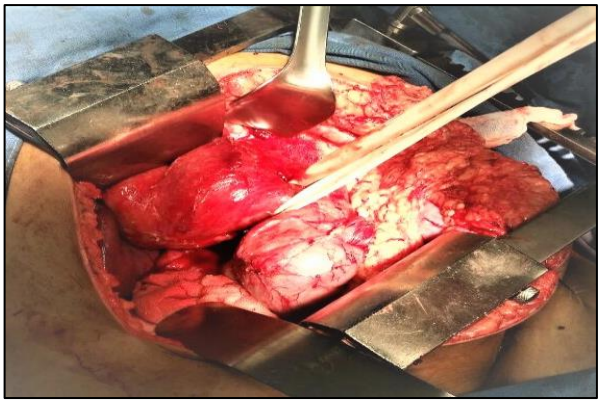

Figure 3: Distal portion of the bile duct referenced with soft sterile drainage.

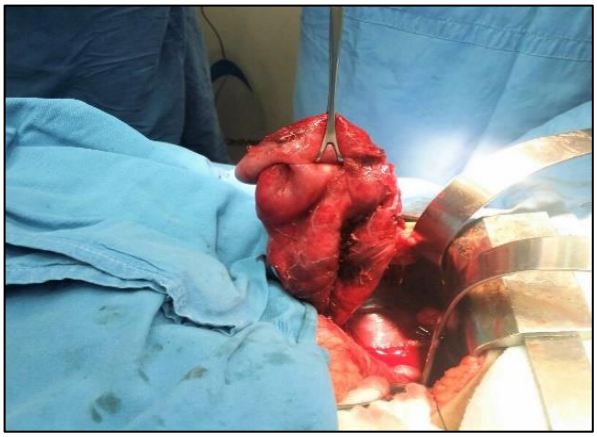

Figure 4: Gentle traction of the cyst once separated from the proximal portion of biliary.

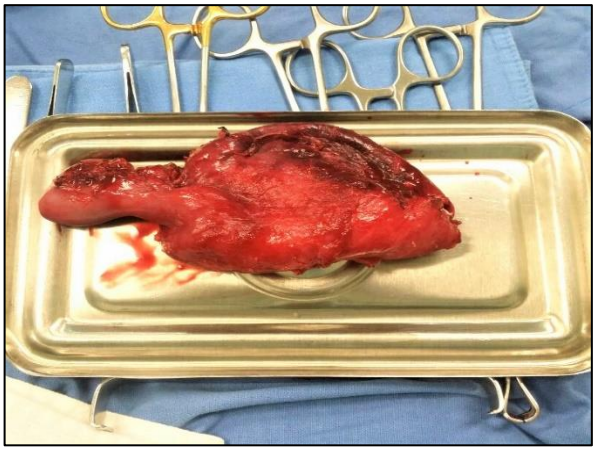

Figure 5: Total surgical sample at the end of the choledochal cyst resection procedure.

In all the previous cases there was administered octreotide postoperative in infusion 600 mcgr per day during five days, until enteral nutrition started, and all patients were sent home at the seventh day without any drainage or complication.

DISCUSSION

Historically, even though the first reported case with a bile duct disorder was by Vater almost 300 years ago, the first well-documented case of a bile duct cyst was in 1852 by Douglas in a 17-year-old woman.[9,10] It is important to take into consideration that despite recent anatomical classifications of choledochal cyst include five forms, type I represents up to 90% of all cases. Today there is a well-established association with the female sex; although there is not much described of the association with pregnancy and biliary cyst.[1,11-13] Even less about the therapeutic management and the role that the surgical techniques and the surgeon play in the resolution of this disease.

Having as a principle that the incidence cystocholedocal disease in pregnancy ranges from 3.3% to 12.2%, it may also have a role in increasing the likelihood of symptomatic biliary tract disease and it appears to increase in the final two trimesters of pregnancy particularly in younger women.[14,15] These malformations occur in two scenarios, cystic dilatation with neonatal presentation or later onset presentation in adult age. There is a possibility that patients present the disease since childhood and alterations in biliary drainage became symptomatic in adulthood or during pregnancy. The postulated mechanisms to explain the cyst spontaneous rupture are abnormal pancreaticobiliary junction leading to efflux of amylase rich fluid into the bile duct, obstruction of the common channel by protein plug and raised intrabdominal pressure. This could be the key point during pregnancy playing a role in the appearance or detection of symptoms.[16] Nevertheless, the exact pathogenesis of this condition is unknown; choledochal cyst in an asymptomatic female can complicate during pregnancy leading to increased fetomaternal morbimortality, especially because the diagnosis and treatment will be delayed and this can lead to pancreatitis, ascending cholangitis, biliary cirrhosis and even cyst rupture with resultant poor fetal and maternal outcomes.[17]

Asymptomatic cysts can accidentally be discovered during pregnancy, with an expansion documented by serial ultrasound examination. Any symptomatic or rapidly enlarging choledochal cyst during pregnancy should be treated immediately. Choledochal cyst presenting with acute cholangitis, must be treated initially with antibiotics, knowing that probably will require cyst decompression to prevent recurrent sepsis.[18] As described in case number two. If it is possible, nonoperative management should be performed until the second trimester or after delivery, when the surgical and anesthetic risks are lower; is important to always take into consideration the risk of rupture during pregnancy may be secondary to the growing uterus with the associated increased intra-abdominal pressure, especially in the intrapartum period during labor, an elective C-section is recommended when the fetus is viable and definitive surgical excision of the cyst should be planned, because there is increased risk of biliary tract carcinoma in non-treated choledochal cysts.[19-21] The definitive treatment of the cyst is necessary for cases treated conservatively during pregnancy; the ideal surgical procedure considered in world literature is complete cyst excision with cholecystectomy followed by biliary reconstruction using a Roux-en-Y hepatic-jejunostomy as the treatment of choice.[22] The di-lemma is still how and when.[23] In this article, according to our experience, we propose it to be four weeks after the resolution of the pregnancy to give more time of effect to pre-operative parenteral nutrition and to be close to the end of the puerperium.

In this series we consider at the principal limitation the small population here presented however it is representative of the population in Mexico and Latin America heterogeneity. The challenges that represent pregnancy young patients and the adaptation of the multidisciplinary team that hospital can offer. A key point that we touch in the approach that has been little explored is the role of nutrition support previous, during and after the definitive surgical treatment, we believe that counteract the intense catabolism inherent to the puerperium taking parenteral nutrition as a strategy, could help to obtain better outcomes as in our series.

CONCLUSION

The treatment of biliary cysts in pregnant women represents a challenge for gynecologists and surgeons. It is important to safeguard the maternal and fetal well-being in each therapeutic decision. In this article we present our proposal on the limitations of conservative treatment such as the presence of cholangitis and fetal suffering, we consider that the surgery treatment it is appropriate four weeks after the resolution of the pregnancy, which, in our opinion, would be appropriate between weeks 35 and 37 of gestation or when viable. The results will be benefited with an adequate parenteral and enteral nutritional preparation, pre and postsurgical due the intense catabolism inherent to the surgery.

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